Infra-hepatic caval resection en bloc with right nephrectomy followed by caval reconstruction for locally advanced caval leiomyosarcoma: A case report and literature review

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Abstract. Retroperitoneal sarcomas often require complex surgical procedures in order to achieve complete resection; in such cases both vascular and visceral resections are needed. When it comes to the need for vascular reconstruction, the type of graft as well as the type of reconstructive process are chosen according to the length and location of the resected segment. Meanwhile, depending on the location of the resected segment, other vascular reconstructions may be needed such as the reimplantation of the renal veins. However, in certain cases, this reimplantation is not mandatory, an adequate renal outflow being reported through the collateral network at this level. We present the case of a 43-year-old patient diagnosed with a large retroperitoneal sarcoma originating from the cava vein invading the right kidney. Resection of the tumor was performed en bloc with caval resection and right nephrectomy, without reimplantation of the left renal vein at the level of the graft. Extended visceral and vascular resections might be needed in order to achieve complete resection of inferior cava vein sarcomas; re-implantation of the left renal vein being not mandatory if rich collateral circulation is present.

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Introduction

Tumoral involvement of the inferior cava vein has been described as being the consequence of the presence of primary lesions at this level such as inferior cava vein leiomyosarcomas or due to the presence of locally advanced urological malignancies such as renal cell carcinoma, adrenal carcinoma or retroperitoneal metastatic adenopathies with urological or gynecological origin invading the inferior cava vein (1,2); in such cases advances in the field of surgical techniques allow performing extended vascular and visceral resections in order to achieve negative resection margins and therefore to offer a chance for cure for these patients (3-9). In the present article, we present the case of a 43-year-old male successfully submitted to surgery for a primitive leiomyosarcoma of the cava vein.

Case report

After obtaining approval of the Ethics Committee of 'Fundeni' Clinical Institute (no. 311/2020), data concerning the patient were reviewed and presented in the present article.

The 43-year-old male with no significant medical history was investigated for diffuse abdominal and dorso-lumbar pain in association with lower limb edema and was diagnosed at the preoperative computed tomography with a large retroperitoneal tumor involving both the cava vein and the right kidney.

After preoperative preparation, the patient was submitted to surgery, the tumor being resected en bloc with infrahepatic, perirenal cava vein resection and right nephrectomy. The continuity of the cava vein was re-established by using a polytetrafluoroethylene (PTFE) prosthesis which was anastomosed with the proximal infra-hepatic cava vein and with the infra-renal cava vein distally. Due to the presence of an adequate collateral network at the level of the left kidney

through both adrenal and gonadal veins, the left renal vein was no longer re-implanted at the level of the synthetic graft (Figs. 1-5).

The postoperative Doppler ultrasound revealed a proper flow at the level of the venous graft while the renal function proved to be an acceptable one; a slight increase in the postoperative values of serum creatinine (at 1.6 mg/dl) in the first postoperative week were encountered. The overall postoperative outcome was favorable, the patient being discharged on postoperative day 13. The anticoagulant injectable treatment consisting of fractioned heparin injections was ended at the time of discharge and replaced with coumadin oral treatment which was administered during the next three months; meanwhile an International Normalized Ratio (INR) value was determined every two weeks, with target values ranging between 2 and 3. At the 3-month follow-up, the patient exhibited a good general condition; the Doppler ultrasound revealed a functional venous graft while the biological tests showed a serum level of creatinine of 1.4 mg/dl. The final histopathological report demonstrated the presence of a caval leiomyosarcoma invading the right kidney; meanwhile negative resection margins were confirmed.

Discussion

Cava vein resection can be imposed by the presence at this level of primary caval leiomyosarcomas, by vascular invasion caused by retroperitoneal sarcomas or other malignant primaries (the most commonly situation being represented by renal tumors) or by metastatic lesions (10). Although initially cava vein invasion was considered as the sign of locally advanced disease and was therefore considered as a contraindication for surgery, improvement of vascular surgical techniques in addition to visceral standard resections have led to the incorporation of such procedures in the therapeutic armamentarium of these cases (3-13). Meanwhile, it has been observed that this aggressive surgical approach remains the only therapeutic option which might increase the overall survival of these patients, especially when it comes to tumors with a primary origin at the level of venous structures such as cava vein leiomyosarcomas (3,12,13). However, the fact should not be omitted that caval resection in the absence of a pre-existent caval obstruction can be hardly tolerated especially in the absence of a well-developed collateral network (14).

Retroperitoneal sarcomas may cause inferior vena cava invasion due to extrinsic involvement, and therefore their resection will impose partial or circumferential venous excision; while in cases in which lateral invasion is present, partial resection followed by primary suture or patch repair could be taken into consideration. Cases presenting circumferential invasion will necessitate circumferential resection; meanwhile, when it comes to primary sarcomas of the cava vein, circumferential resection is usually the option of choice (15).

In this respect, it should not be overlooked that caval resection may increase the cardiac preload, may increase the risk of venous congestion and the peripheral venous pressure resulting in lower extremity edema and deep venous thrombosis (16).

Depending on the length of the resected segment and on the presence and patency of collateral circulation, various methods of reconstruction might be taken into consideration. The presence of collateral circulation, if patent, may allow performing caval ligation without further reconstruction; moreover, interruption of the renal veins at the time of resection might be well tolerated if an adequate collateral venous return is provided through the adrenal veins. However, attention should be focused at the time of resection in order not to destroy the network of collaterals which is expected to provide an adequate venous return (3,10,12,13,15,17). Moreover, this aspect is particularly important in the case of the left kidney, in which the left gonadal and adrenal veins appear to play a crucial role in providing an adequate venous return, as for the right kidney, the absence of these collaterals might pose significant issues in terms of venous return (15).

According to the length of resection of the cava vein, various types of reconstruction have been proposed, ranging from primary repair, patch placement or segmental resection followed or not by venous reconstruction; as mentioned before, cases in which an adequate collateral circulation is present may be candidates for solely resection and no further reconstruction (10).

When it comes to the types of materials which can be used for venous replacement, both synthetic and natural grafts have been proposed. Therefore, using a circular polytetrafluoroethylene (PTFE) prosthesis may be the option of choice due to the fact that in a significant number of cases, it is more facile to be obtained. If this is the option of choice, attention should be paid to the diameter of the graft; the general recommendations underlining the fact that a lower diameter prosthesis is more efficient due to the fact that it seems to provide a faster velocity at its level (15,18). In cases in which an autologous graft is available, the superficial femoral vein has been widely used; meanwhile, cryopreserved grafts can be also used (15). As for the details of surgical technique, it is considered that the reconstruction should proceed from distal to proximal; meanwhile the bifurcation of the caval vein should be preserved as much as possible in order to make more facile the distal reconstruction (15). In the meantime, if the perirenal segment is resected, it is recommended to perform first the distal and proximal anastomoses of the graft followed by renal vein reimplantation if possible; if the both renal veins are to be implanted, the right one should be first reinserted due to the lack of collaterals of the right kidney when compared to the left kidney (15,19-21). As for the anastomosis at the level of the proximal end, the most common site of this anastomosis is the infra-hepatic area; in certain cases in which the tumor also involves this segment, a retrohepatic resection and anastomosis are required. However, in such cases, attention should be focused on the risk of destroying the venous branches of the caudate lobe, which can cause significant bleeding (15).

Furthermore, it should not be overlooked that the only chance for cure in such cases is represented by the achievement of negative resection margins (22,23), and therefore the length of the resected segment should be long enough in order to achieve this desiderate (24). Moreover, it has been demonstrated that administration of adjuvant treatment can be difficult to be administered due to the high sensitivity of the healthy tissues around the field of resection (25). The utility of negative resection margins is also sustained by the observation that 77% of the sarcoma-related deaths are caused by

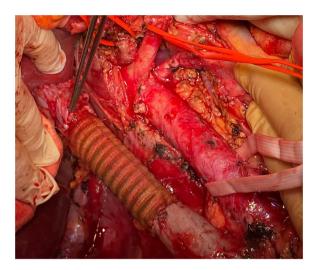


Figure 1. The final aspect after cava vein replacement; anastomosis of the graft with the infra-hepatic portion of the cava vein.

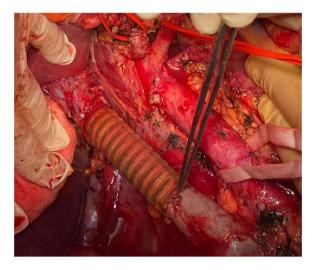


Figure 2. The final aspect after cava vein replacement; anastomosis of the graft with the graft of distal, infra-renal portion of the cava vein.

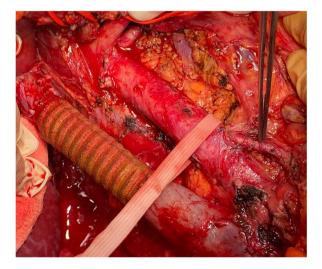


Figure 3. Arterial branches of the abdominal aorta were entirely preserved; however the left renal vein was no longer reinserted at the level of the cava vein, with adequate perfusion of the left kidney being maintained through collateral circulation.

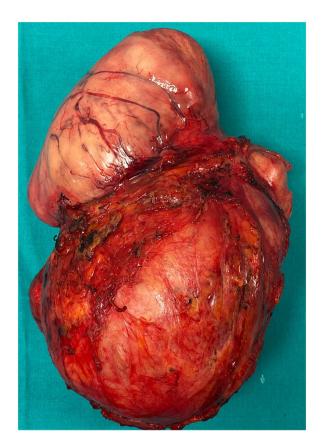


Figure 4. The specimen: retroperitoneal sarcoma originating from the cava vein invading the right kidney.

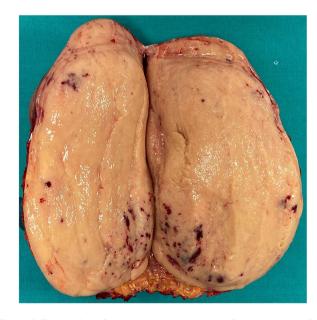


Figure 5. Transection of the specimen revealed the fleshy structure of the tumor; typical aspect for a sarcoma.

the development of local recurrence in the absence of distal metastases (26).

One of the largest studies which analyzed the effectiveness and safety of cava vein resection as part of extended oncological procedures was conducted by Ruiz *et al* (10); the study included 52 patients submitted to cava vein resection

as part of various oncological procedures; among these cases there were 5 patients diagnosed with primary cava vein leiomyosarcomas, 11 renal cell carcinomas, 7 testicular carcinomas, 5 cholangiocarcinomas, 10 retroperitoneal sarcomas and a variety of other histopathological types and subtypes of lesions. As for the option of choice for reconstruction, in 17 cases, primary repair was the option of choice; in the other 18 cases, a patch angioplasty was required while in the remaining 17 cases, graft interposition was the option of choice. Among the latter category, PTFE grafts were used in 13 cases, Dacron grafts were preferred in 2 cases while in the remaining 3 cases homologous grafts were used; the authors decided to reinsert the left renal vein at the level of the prosthesis in 3 cases. Postoperatively the overall complication rate was 75%, 10 cases necessitating reoperation; however, among cases in which graft interposition was performed only 2 cases developed postoperative graft thrombosis and secondary lower limb lymphedema. Meanwhile the authors underlined the fact that graft thrombosis was significantly higher among cases in which non-ringed grafts were used as well as among cases in which the diameter of the graft was wider than 18 mm. As for the long-term outcomes, the authors reported a 2-year survival rate of 64.7% and a 2-year patency rate of 77.5% demonstrating in this way the effectiveness and safety of the method (10).

In conclusion, retroperitoneal sarcomas originating from the cava vein might require extensive resection followed by demanding reconstruction of the venous contiguity in order to re-establish a functional venous outflow at this level. In cases in which circumferential resections are needed, the reconstruction can be performed by allograft or autologous grafts. In such cases, a debatable subject is related to the necessity of performing a reimplantation of the renal veins. The presence of an adequate collateral network seems to provide an acceptable venous return especially for the left kidney; the most important collateral venous drainage pathways being represented by the left adrenal and gonadal vein.

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Availability of data and materials

Further information regarding the case presentation is available upon request.

Authors' contributions

NB contributed to the conception of the study, collected, analyzed and interpreted the data from the literature and critically revised the manuscript. IoB contributed to the conception of the study, performed the literature research, drafted the manuscript and was responsible for confirming the authenticity of all the raw data. VB contributed to the conception of the study, performed the literature research, drafted the manuscript and was responsible for confirming the

authenticity of all the raw data. IrB contributed to the interpretation of the data from the literature, collected, analyzed and interpreted the data corresponding to the patient and critically revised the manuscript. IC collected, analyzed and interpreted the data corresponding to the patient and critically revised the manuscript. All authors read and approved the final manuscript for publication.

Ethics approval and consent to participate

The Ethics Committee of 'Fundeni' Clinical Institute (Bucharest, Romania) (no. 311/2020) approved the study.

Patient consent for publication

Patient consent for publication was obtained and signed by the patient on 23/08/2020.

Competing interests

There are no competing interests to declare regarding this study.

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